Cesarean Section in a Parturient with Carpenter’s Syndrome and Corrected Tetralogy of Fallot

Abstract Type: Case Report/Case Series
Manish Bhardwaj, M.D., FRCA; Edwin Bone, FRCA; Caroline Grange, FRCA
Oxford Radcliffe Hospitals NHS Trust

Introduction: Carpenter’s syndrome is a rare (1 in one million) autosomal recessive disorder characterized by craniofacial malformations, syndactyly, heart defects & obesity. We report the successful perioperative management of a parturient with Carpenter’s syndrome with deteriorating cardiac function.

Case: A nulliparous, super obese (BMI 51) 22 year old parturient with Carpenter’s syndrome (tetralogy of Fallot and craniosynostosis had been corrected at 10 months and 7 years of age respectively) was referred to our centre at 22 weeks gestation with increasing breathlessness. Additional problems included difficult intravenous access, needle phobia and osteoarthritis (OA). Although exercise tolerance was limited due to OA, she complained of increasing breathlessness and worsening palpitations from 20 weeks gestation. ECG revealed sinus rhythm and RBBB and initial echocardiogram showed good left and right ventricular (LV, RV) function with mild pulmonary regurgitation (PR). A multidisciplinary team (obstetrician, cardiologist, anesthesiologist, and neonatologist) was involved in her management. Diuretic and low molecular weight heparin (LMWH) were started. At 35 weeks gestation she was admitted with worsening pulmonary oedema. Echocardiogram showed moderately impaired LV and RV function, severe RV hypertrophy / dilatation and severe PR. The decision to deliver the fetus was made due to maternal concerns despite further diuresis and fluid restriction. Airway assessment was normal and the LMWH was stopped 12hrs prior to surgery. Dexamethasone was used to improve fetal pulmonary maturation and antacid prophylaxis given. Despite her needle phobia, the patient opted for regional anesthesia. Peripheral, arterial and internal jugular cannulae were inserted (with much persuasion due to her needle phobia) prior to anesthesia. Following antacid prophylaxis, a low dose combined spinal epidural (spinal dose 1.2mls 0.5% heavy bupivacaine) with careful epidural top up were used to achieve T4 to S5 block. A healthy female baby (2.24 kg, Apgars 9 & 10 at 1 and 5 mins respectively) was delivered. After delivery, an infusion of oxytocin was started and antibiotics given. The patient remained hemodynamically stable throughout surgery (estimated blood loss 800 mls). Following surgery she was transferred back to the cardiology ward for monitoring and LMWH was recommenced. She was discharged home with her baby on the 10th post-operative day.

Discussion: Multidisciplinary care, frequent thorough antenatal assessments, optimization of worsening maternal cardiac symptoms, invasive monitoring, the use of a titratable / cardiovascular stable anesthetic technique and consideration of her other problems (morbid obesity and needle phobia) were the key components in the successful management of this rare and challenging condition.

Reference: